Appl. No. TO BE ASSIGNED Amdt. dated June 30, 2003 Preliminary Amendment

This listing of claims will replace all prior versions, and listings of claims in the application:

## Listing of Claims:

1. (original) A method of treating a patient with Pompe's disease, comprising: administering to the patient a therapeutically effective amount of human acid alpha glucosidase.

## 2-34. (canceled)

- 35. (new) A method of treating a human patient with infantile Pompe's disease, comprising: administering to the patient at least 10 mg/kg body weight per week of human acid alpha glucosidase, whereby the patient survives to be at least one year old.
- 36. (new) The method of claim 35, wherein the patient is administered at least 60 mg/kg body weight per week.
- 37. (new) The method of claim 35, wherein the patient is administered at least 120 mg/kg body weight per week.
- 38. (new) The method of claims 35, wherein the patient is administered a single dosage of alpha-glucosidase per week.
- 39. (new) The method of claim 35, wherein the patient is administered three dosages of alpha-glucosidase per week.
- 40. (new) The method of claim 35, wherein the amount is administered per week for a period of at least 24 weeks.
- 41. (new) The method of claim 35, wherein the alpha-glucosidase is administered intravenously.
- 42. (new) The method of claim 35, wherein the alpha-glucosidase was produced in milk of a transgenic mammal.

- 43. (new) The method of claim 35, wherein the alpha-glucosidase is predominantly in a 110 kD form.
- 44. (new) The method of claim 35, further comprising monitoring a level of human acid alpha glucosidase in the patient.
- 45. (new) The method of claim 44, further comprising administering a second dosage of human acid alpha glucosidase if the level of alpha-glucosidase falls below a threshold value in the patient.
- 46. (new) The method of claim 35, wherein the human alpha glucosidase is administered intravenously and the rate of administration increases during the period of administration.
- 47. (new) The method of claim 46, wherein the rate of administration increases by at least a factor of ten during the period of administration.
- 48. (new) The method of claim 46, wherein the rate of administration increases by at least a factor of ten within a period of five hours.
- 49. (new) The method of claim 46, wherein the patient is administered a series of at least four dosages, each dosage at a higher strength than the previous dosage.
- 50. (new) The method of claim 49, wherein the dosages are a first dosage of 0.03-3 mg/kg/hr, a second dosage of 0.3-12 mg/kg/hr, a third dosage of 1-30 mg/kg/hr and a fourth dosage of 2-60 mg/kg/hr.
- 51. (new) The method of claim 50, wherein the dosages are a first dosage of 0.1-1 mg/kg/hr, a second dosage of 1-4 mg/kg/hr, a third dosage of 3-10 mg/kg/hr and a fourth dosage of 6-20 mg/kg/hr.

- 52. (new) The method of claim 51, wherein the dosages are a first dosage of 0.25-4 mg/kg/hr, a second dosage of 0.9-1.4 mg/kg/hr, a third dosage of 3.6-5.7 mg/kg/hr and a fourth dosage of 7.2-11.3 mg/kg/hr.
- 53. (new) The method of claim 52, wherein the dosages are a first dosage of 0.3 mg/kg/hr, a second dosage of 1 mg/kg/hr, a third dosage of 4 mg/kg/hr and a fourth dosage of 12 mg/kg/hr.
- 54. (new) The method of claim 49, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.
- 55. (new) The method of claim 49, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1 hr, 0.5 hr and 3 hr respectively.
- 56. (new) The method of claim 35, wherein the dosage is at least 40 mg/kg body weight per week.
- 57. (new) The method of claim 35, further comprising detecting a reduction in hypertrophic cardiomyopathy responsive to the administering.
- 58. (new) The method of claim 35, further comprising detecting increased alpha-glucosidase activity in muscle responsive to the administering.
- 59. (new) The method of claim 35, wherein the human alpha-glucosidase is administered to a population of patients with infantile Pompe's disease and at least 50% survive more than six months post diagnosis of the infantile Pompe's disease without life-saving intervention.
- 60. (new) A method of treating a human patient with infantile Pompe's disease, comprising: administering to the patient at least 15 mg/kg body weight per week of human acid alpha-glucosidase.

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- 61. (new) The method of claim 60, wherein the patient is administered at least 20 mg/kg body weight per week of human acid alpha-glucosidase.
- 62. (new) The method of claim 60, wherein as a result of the treatment method the patient survives to be at least 1 year old.
- 63. (new) The method of claim 62, wherein the acid alpha-glucosidase is administered intravenously once a week.
- 64. (new) The method of claim 62, wherein the acid alpha-glucosidase is administered intravenously twice a week.